

Jaccoud's arthropathy: a rare but well-known clinical entity

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Jaccoud arthropathy is a deforming non-erosive arthropathy characterized by ulnar deviation of the 2nd to 5th fingers with metacarpophalangeal joint subluxation that is correctable with physical manipulation [Mittermayer S, Viviane M. Jaccoud's arthropathy. *N Engl J Med* 2015;373:e1.]. It was traditionally described as occurring post-rheumatic fever but also seen in association with systemic lupus erythematosus, psoriatic arthritis, inflammatory bowel disease, and malignancy [Palazzi C, D'Amico E, De Santis D, Petricca A. Jaccoud's arthropathy of the hands as a complication of pyrophosphate arthropathy. *Rheumatology (Oxford)* 2001;40:354–355.]. It is thought to be related to ligamentous laxity. It typically affects the metacarpophalangeal joints but can also affect the proximal interphalangeal joints of the hands, wrists, and knees [Spina MF, Beretta L, Masciocchi M, Scorza R. Clinical and radiological picture of Jaccoud arthropathy in the context of systemic sclerosis. *Ann Rheum Dis* 2008;67:728–729.]. Hand radiographs typically show marked ulnar subluxation and deviation at the metacarpophalangeal joints with absence of erosions. We present a case in a very young female with no prior history of rheumatic fever or acute arthritis at any stage of illness.

Keywords

Jaccoud's arthropathy • Rheumatic heart disease • Deforming arthritis

Learning points

- Jaccoud's arthropathy is a benign chronic arthropathy without functional impairment and needs to be differentiated from fixed and non-correctable deformities of rheumatoid arthritis.
- Also with the help of proper history and systematic examination (especially careful auscultation), we can identify the potential cause of arthropathy like Rheumatic heart disease, Systemic lupus erythematosus, etc.

Patient presentation

A 15-year-old female child presented to us with complaint of fatigue on exertion. No fever/shortness of breath/chest pain/palpitation/cyanosis/syncope. There was no history of fever with joint pain or swelling suggestive of acute rheumatic fever in the past. There were no symptoms of congestive heart failure. On examination, she was alert and oriented. Blood pressure = 100/70 mmHg, heart rate =

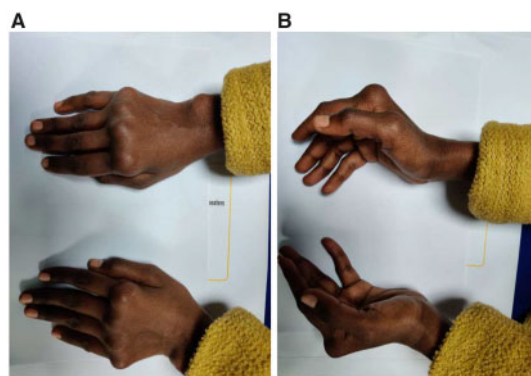


Figure 1 (A and B) Photograph of both hands of our patient showing flexion deformities and ulnar deviation at the metacarpophalangeal and hyperextension deformity at the proximal interphalangeal joints.

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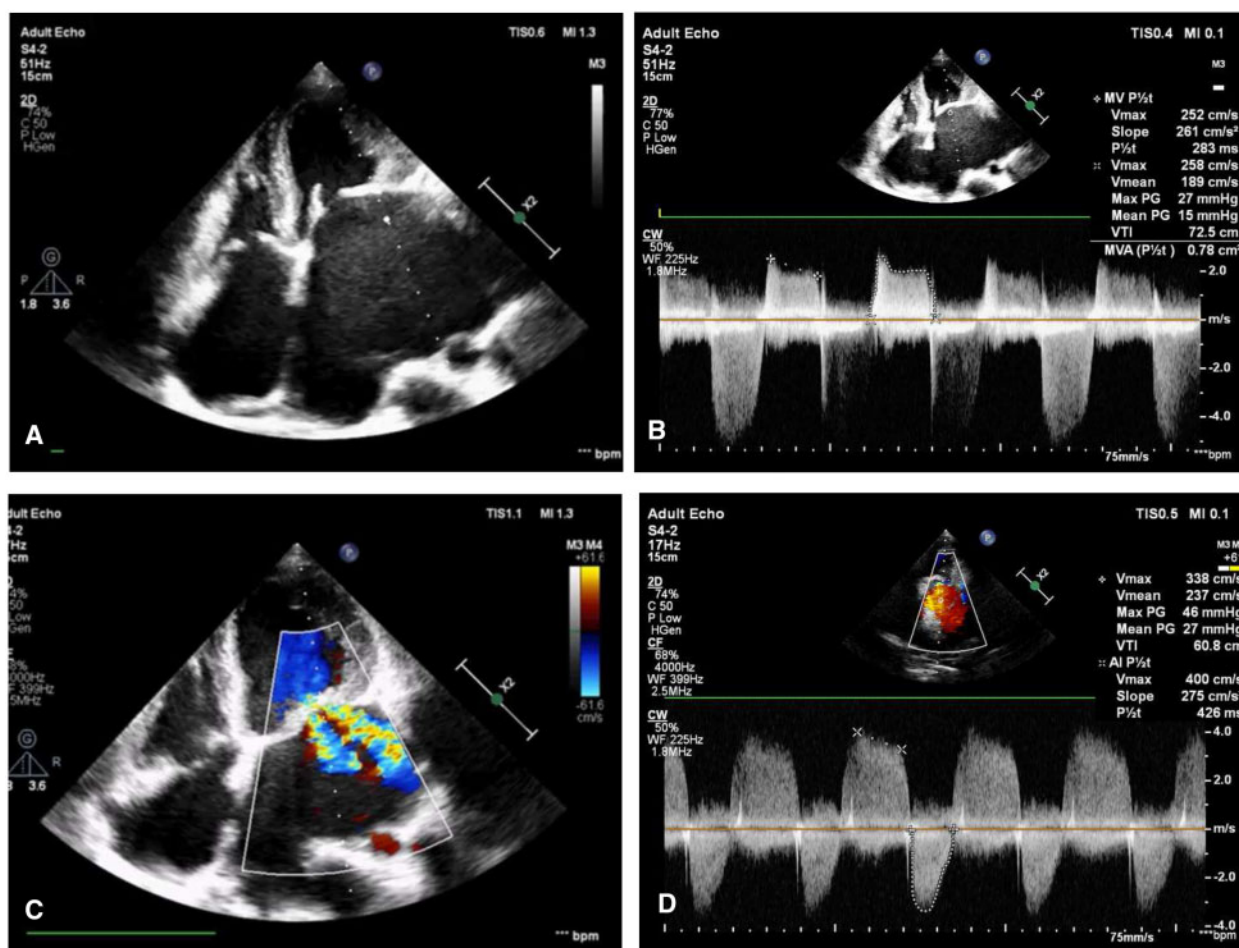


Figure 2 (A) Two-dimensional echocardiography in apical four chamber view showing anterior mitral leaflet thickened with domed shape and posterior mitral leaflet thickened and fixed with giant left atrium. (B) Doppler at the level of mitral valve in apical four chamber view showing mitral valve area by pressure half time is 0.78 cm² with mean gradient of 15 mmHg with moderate mitral regurgitation. (C) Colour Doppler across mitral valve in apical four chamber view showing moderate mitral regurgitation. (D) Doppler at level of aortic valve in apical five chamber view showing mild aortic stenosis with mean gradient of 27 mmHg and mild aortic regurgitation with pressure half time of 400 cm/s.

90/min, and saturation = 99% on room air. On cardiovascular examination, there was a low pitched mid-diastolic rumbling murmur and a high pitched, soft blowing pansystolic murmur at the mitral area. At the aortic area, harsh mid-systolic ejection murmur radiating towards carotid is heard. At neo-aortic area, high pitched, soft blowing, early diastolic decrescendo murmur is heard. Examinations of major joints did not reveal any sign of acute rheumatic activity. However, her both hands showed fusiform swelling of the metacarpal-phalangeal joints with flexion deformity and ulnar deviation which was correctable. The palmar fascia was not involved. There was also a hyper-extension deformity of the proximal interphalangeal joints (Figure 1). The patient was not troubled by these deformities and on questioning said that gradual increase had occurred in these deformities in the past 2 years. There were no features suggestive of Marfan's syndrome or other heritable connective tissue disorder.

Initial work up

Investigations for rheumatic activity yielded negative results. Tests for antinuclear antibody and rheumatoid factors were also negative. Her electrocardiogram showed left ventricular hypertrophy. On 2D echocardiography, in parasternal long-axis view, anterior mitral leaflet was thickened with diastolic doming and posterior mitral leaflet was thickened and fixed suggestive of Rheumatic heart disease. Mitral valve area by planimetry was 0.9 cm² and by pressure half time was 0.78 cm² indicating severe mitral stenosis. Other findings include moderate mitral regurgitation, mild aortic stenosis, mild aortic regurgitation, moderate tricuspid regurgitation, severe pulmonary artery hypertension, moderate pulmonary regurgitation with normal left ventricular (LV)/right ventricular (RV) function (Figure 2).



Figure 3 Radiograph of both hands of our patient showing flexion deformities and ulnar deviation at the metacarpophalangeal and hyperextension deformity at the proximal interphalangeal joints. There was no evidence of any erosion in the articular surface of any joint.

Diagnosis and management

Her hand radiograph showed marked ulnar subluxation and deviation at the metacarpophalangeal joints with absence of erosions (Figure 3). Based on history, examinations, X-ray, and echocardiography, she was diagnosed to have Rheumatic heart disease, severe mitral stenosis, moderate mitral regurgitation, mild aortic stenosis, mild aortic regurgitation, moderate tricuspid regurgitation, severe pulmonary artery hypertension, moderate pulmonary regurgitation with normal LV/RV function with Jaccoud's arthropathy. She was started on injection Benzathine Penicillin prophylaxis, oral beta-blocker, low-dose diuretics with physiotherapy of hand.

Follow-up

During follow-up, her symptoms improved. She was continued on injection Benzathine Penicillin prophylaxis and physiotherapy of hand with plan for mitral valve replacement.

Conclusion(s)

Jaccoud's arthropathy is not commonly seen in Western world. It has got clinical significance—as presence of this arthropathy mandates further investigation to identify a potential cause—rheumatic, SLE, and other diffuse connective tissue diseases. This arthropathy is potentially correctable.

Lead author biography



Dr Ashish Kumar Golwara obtained his DNB Cardiology degree from National Board of Examinations, Delhi, India in 2019. He is currently working as consultant interventional cardiologist in Medica Heart Institute, Patna, Bihar, India.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

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